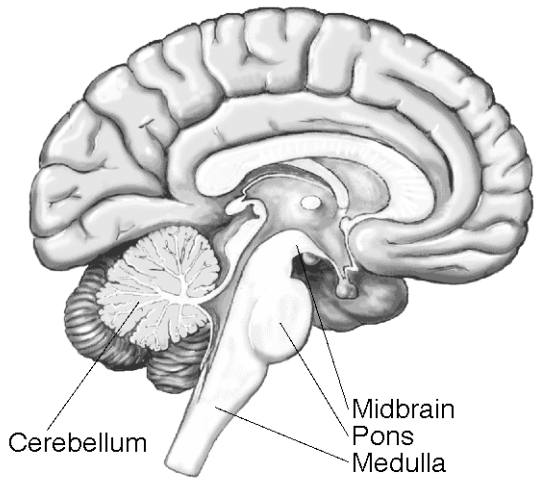
Arnold-Chiari Malformation

Thomas Farley, MA
Research and Statistics Manager
Arkansas Spinal Cord Commission
August 2005

Arnold-Chiari Malformation (ACM) is the abnormally (small) development of the lower, back part of the skull. As a result, there is less room for the parts of the brain housed in this area. The midbrain, pons, medulla and cerebellum are crowded together often with the cerebellum pushing down into the spinal cord column. This crowding interferes with the normal operation of these parts of the brain and/or spinal cord. In addition, the movement of cerebrospinal fluid may be blocked creating increased pressure in the brain and/or around the spinal cord.

Cerebrospinal fluid (CSF) is created in the brain cavity and bathes the surfaces of the brain and spinal cord. The fluid acts as a shock absorber and is necessary for proper functioning. The fluid is absorbed by the body at the base of the spinal cord.



ACM was named for Hans Chiari, an Austrian pathologist who described the condition in the late 1800's. Three years later Julius Arnold, a German pathologist, described the same condition; thus, the malformation was later referred to as Arnold-Chiari.

Alternate names for ACM are cerebellomedullary malformation syndrome, Arnold-Chiari deformity, Arnold-Chiari syndrome, Chiari malformation and Chiari.

Types of ACM

There are three types of ACM:

ACM I is characterized by protrusion of the brain tissue below the opening of the base of the skull and may include an abnormal fluid blister-like cavity (syringomyelia) and larger than normal accumulations of CSF in the skull (hydrocephalus).

ACM II is a protrusion of both the cerebellar brain tissue and the brain stem through the hole in the skull. This form of the malformation may be accompanied by syringomyelia and hydrocephalus, but, in addition, it is often associated with spina bifida (spina bifida is a congenital disorder in which the vertebrae fail to completely encircle the spinal cord).

ACM III includes all of the characteristics listed in 1 and 2 plus a bulge of the spinal cord caused by a sac or balloon-like abnormality (myelomeningo-

cele) that may contain CSF, and/or issue from the spinal cord membranes.

Symptoms

The cerebellum transmits information to the body about maintenance of equilibrium and balance, regulation of muscle tension and coordination of limb control. Compression of the cerebellum and surrounding tissues is likely to result in: headaches, dizziness, neck pain, tiredness, stiffness, vomiting, difficulty swallowing, gagging, muscle weakness in head and face, discomfort in the arms and legs and various degrees of mental impairment.

Signs and symptoms of ACM usually are noticeable shortly after birth. If symptoms do not appear in childhood, persons with ACM may show signs of worsening brain impairment as a teenager or adult. This often presents as involuntary, rapid downward eye movements, also dizziness, headache, double vision, ringing in the ears, inability to coordinate movements and sudden pain around the eyes.

Causes, Prevalence and Risk Factors

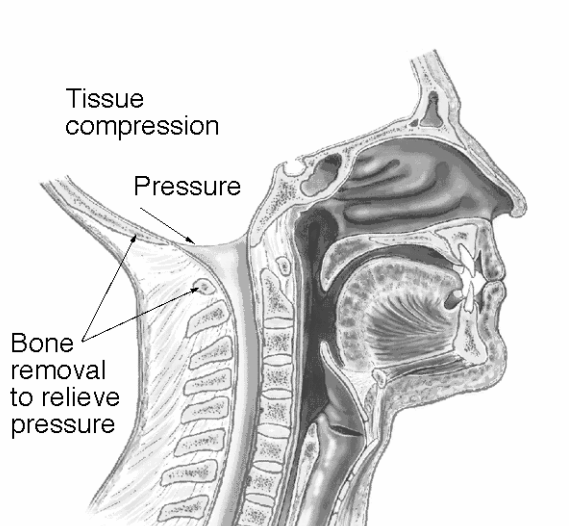
The exact cause of ACM is not known. As mentioned previously, it is known that the back, bottom part of the skull is abnormally developed, resulting in a smaller space to hold the cerebellum and other brain parts and tissues. However, what causes the abnormal development of the skull is unknown. Some researchers suspect that this condition is inherited from the individual's parents but convincing evidence is lacking.

Prevalence and incidence rates of ACM are difficult to estimate because ACM is not a reportable condition and because population-based studies have not been conducted to determine the actual rates of occurrence. ACM is not a common condition. Although it is considered primarily a congenital abnormality it can be acquired later in life through accident or by other unknown means. The Chiari Institute in Great Neck, New York estimates that there are 200,000 to 2 million Americans with ACM and that around 3,500 Chiari operations are performed each year in the United States. ACM type II is associated with spina bifida which has an incident rate of approximately 2 per 1,000 live births.

More females have ACM than males. Estimates range up to three times as many females as males have ACM. Other risk factors are unknown.

Diagnosis

ACM is often misdiagnosed as a variety of other conditions and diseases. Only a Computed Tomography (CT) scan or preferably a Magnetic Resonance Imaging (MRI) scan can confirm that ACM exists. An MRI scan is preferred because of its ability to capture a clear image of the soft tissues of the body, in this case the cerebellum and other brain and spine tissues.



Treatment

Treatment options are very limited. Only surgery can enlarge the area and allow more room for the cerebellum and other tissues. This is done by removing part of the lower skull at the back of the head and perhaps by removing some of the upper portions of the vertebra in the spina column. How much is removed can not be determined until a thorough examination has been conducted by a neurosurgeon experienced in the Chiari condition.

All surgeries have risks and this particular surgery is serious and should not be undertaken lightly. Often more than one surgery may be required. If symptoms are manageable, surgery may not be indicated. However, if symptoms become unbearable or the person gradually deteriorates mentally or pressure in the head or spinal cord increases, surgery, at present, is the only option.

Prognosis - Living with ACM

Generally speaking, the more severe the symptoms then the worse the prognosis. The signs and symptoms of most individuals will improve with surgery and many will have minimal symptoms. Others may have to have other surgical procedures. A severe case of ACM in an infant can be life threatening.

More Information

More information regarding Arnold-Chiari Malformation may be obtained from the:

World Arnold-Chiari Malformation Association **www.pressenter.com/~wacma/**

Spina Bifida Association of America **www.sbaa.org/site/PageServer?pagename=ABS_chiari**

National Institute of Neurological Disorders and Stroke **www.ninds.nih.gov/disorders/chiari/chiari.htm**

References

FMS Canberra. (2005). Retrieved August 15, 2005, from http://fmscanberra.tripod.com/acm.htm

MedFriendly. (2005). Retrieved August 15, 2005, from http://www.medfrendly.com/chiarimalformation.html#look

National Organization for Rare Disorders. (2005). Retrieved from http://www.peacehealth.org/kbase/nord/nord85.htm

The Chiari Institute. (2205). http://www.chiariinstitute.com/chiari_about.html

World Arnold-Chiari Malformation Association. (2005). Retrieved August 15, 2005, from http://www.pressenter.com/~wacma/